

Case Report

Poland Syndrome: Case Report and Discussion

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Case Report

A 25-year-old female presented with a history of breast asymmetry since puberty. She denied any significant past medical or surgical history. She took no medications, had no known drug allergies and neither smoked tobacco nor drank alcohol. She denied any breast pain or nipple discharge. On physical examination, the patient had asymmetry of the chest wall with the left breast being significantly larger than the right (Figure 1).



Figure 1: Pre-operative photograph.

The nipple/areola position on the left was superior to that on the right and there were no superficial skin changes. She had full range of motion of the upper extremities and no anomalies of the hands or fingers. There was no palpable adenopathy. A staged approach was chosen for the breast reconstruction. At the first procedure, a supraperiosteal plane was developed for placement of a tissue expander. Of note, there was no pectoralis major muscle under which an implant is typically placed (Figure 2).

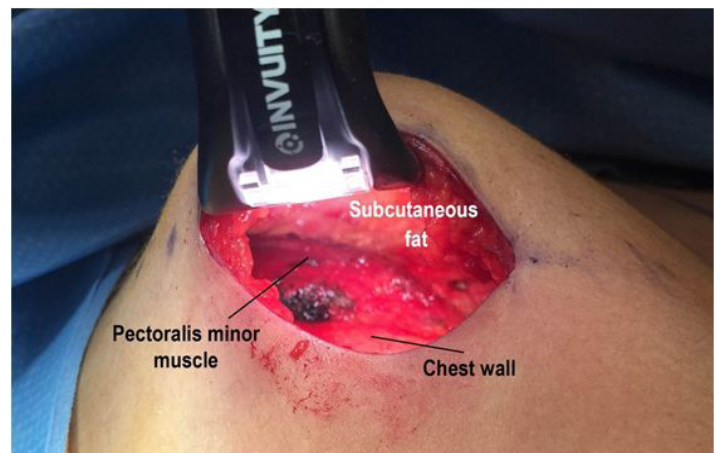


Figure 2: Operative view of the chest wall via an inframammary incision.

The expander was later exchanged for a more permanent implant, followed by a contralateral mastopexy to improve overall symmetry. Possible etiologies to breast asymmetry include hormonal changes or traumatic injuries. At times, the exact cause of asymmetric chest wall is unknown, but occasionally, an underlying medical or skeletal condition may cause asymmetry. Juvenile (or virginal) hypertrophy of the breast is a rare condition in which one breast grows significantly larger than the other. It presents with obvious asymmetry and psychological concerns for the patient. It is typically treated with surgery. Malignancy must always be considered in patients with breast concerns but will not cause hypoplastic changes on the affected side. Other possible causes include scoliosis, or curvature of the spine, and deformities in the chest wall.

Introduction

Poland syndrome is a congenital disorder in which affected individuals most notably lack a portion of the pectoralis major muscle. It is a rare congenital malformation first noted by Lallemand in 1826 [1]. The eponym was assigned to Alfred Poland, a medical student and anatomist, who later described the condition in 1841

[2]. The syndrome is invariably unilateral, with a higher incidence in female than male patients. The right side is more commonly affected than the left. It is associated with various degrees of anomalies of the chest wall and upper extremity. Hypoplasia of the breast may be noted during pubertal development. The nipple-areolar complex may vary in position, color, and/or size. Most cases of Poland syndrome are sporadic, and the etiology remains uncertain [3]. It has been proposed that an alteration of blood flow through the subclavian artery on the affected side leads to the growth disturbance. This diminution of blood flow may be due to rapid growth of the ribs on the affected side compressing the arteries and or veins supplying the pectoralis muscle. The diminution may also be due to a congenital malformation of the embryonic blood vessel serving the muscle.

The condition is seldom familial. Similar defects have been noted with exposure to certain drugs, such as thalidomide. Poland syndrome is frequently associated with Hodgkin disease, leukemia, leiomyosarcoma, renal tumors, or lung tumors [4-6]. There are a few reports on the association between Poland syndrome and breast cancer. One review highlighted seven papers reporting seven patients with Poland syndrome and breast cancer [7].

Clinical Manifestations

The hallmark of Poland syndrome is involvement of the chest musculoskeletal structures including agenesis of the sternocostal head of the pectoralis major muscle and/or absence of costal cartilages or ribs 2, 3, and 4, or 3, 4, and 5. The latter may be associated with herniation of lung tissue [8]. The result of not having this portion of muscle is asymmetry, notable for an irregular contour of the ipsilateral chest with an absent anterior axillary fold. Involvement of the upper extremity may include syndactyly and/or brachydactyly. Specifically, this is characterized by unilateral, simple syndactyly with ipsilateral limb hypoplasia and pectoralis muscle agenesis.

Management

Management of Poland syndrome differs depending on the symptoms but may include reconstruction by placement of an implant and/or transfer of the latissimus dorsi muscle. The goals of treatment are to address the three areas of concern - hypoplasia of the breast, deficiency of soft tissue, and the bony deformity (if necessary). Patients may appear at any age or stage of development. Therefore, treatment must be tailored to the individual and may require stages. Early reconstruction of the chest wall anomaly may be beneficial in the younger patient who presents prior to puberty. Techniques to address the chest wall deformity include pectus repair (if present), placement of alloplastic material, or addition of autologous tissue. The latter may involve serial lipoaugmentation from available donor sites with or without an alloplastic implant [9,10]. Autologous rib grafts have been recommended to stabilize the skeletal defect along with a Dacron patch to replace the

absent fascia and provide a base for soft-tissue repair [11]. Due to concern for the alloplastic prosthesis and incomplete cosmetic improvement, reconstruction with a pedicled, latissimus muscle flap has been used.

In females, addressing the reconstruction of the breast in two stages may be advantageous. This might commence with placement of an ipsilateral tissue expander, followed by a pedicled muscle flap, with or without a more permanent implant. The latissimus dorsi muscle is most often used, on account of its proximity, size and favorable rotation arc. Use of a free, contralateral latissimus dorsi muscle flap has been described in a 2-year-old patient [12]. Immediate reconstruction of the breast may be preferred in the older patient after breast development is complete [13]. Again, a combination of tissue expansion followed by implant and/or muscle flap reconstruction is preferable. In males, preoperative imaging can be used to fabricate a custom silicone implant, which may be preferable since it spares the use of donor muscle(s). Post-operatively, any residual contour irregularities can be addressed with lipoaugmentation.

Operative management of syndactyly in Poland syndrome is determined by the severity of hand involvement and the resulting anatomical dysfunction [14]. Reconstruction of the fused digit is recommended in most cases of Poland syndrome since patients have notable brachydactyly, and digital separation can improve functional length. Any fusions involving border digits should be managed early to address length discrepancies and the resultant distortion of the affected digits. Middle digits can be corrected secondarily.

Outcomes following correction of the chest wall and breast deformity in Poland syndrome have generally been good. The extent of involvement varies widely and a variety of techniques are required to correct them. It is important to be versatile in numerous techniques to address the various manifestations of the syndrome. Long-term results following correction of the breast/chest wall anomaly in a cohort of patients with Poland syndrome was described by Seyfer, et al [15]. The majority of women underwent reconstruction with soft prosthetic devices, with or without a latissimus dorsi flap. Custom-made implants, on the other hand, required premature removal in 75 percent. Contralateral procedures, namely mastopexy, for balance and symmetry were necessary in 62 percent.

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